HYPERTROPHIC PYLORIC STENOSIS COMPLICATING ESOPHAGEAL ATRESIA WITH TRACHEOESOPHAGEAL FISTULA

T. Dağlı, M.D.* / G. Kıyan, M.D.**

* *Assistant Profesor, Department of Pediatric Surgery, Faculty of Medicine, Marmara University, Istanbul, Turkey.*
** *Research Assistant, Department of Pediatric Surgery, Faculty of Medicine, Marmara University, Istanbul, Turkey.*

SUMMARY
In this paper a case of hypertrophic pyloric stenosis (HPS) complicating esophageal atresia (EA) and tracheoesophageal fistula (TEF) is presented. Being a rare but an important anomaly, the association of hypertrophic pyloric stenosis with esophageal atresia and tracheoesophageal fistula is discussed emphasizing the early diagnostic problem.

Key Words: Hypertrophic Pyloric Stenosis, Esophageal Atresia, Tracheoesophageal Fistula.

INTRODUCTION
The incidence of associated congenital anomalies in patients with esophageal atresia, (EA) and tracheoesophageal fistula (TEF) is about 50 percent (1,2). The frequency of anomalies of gastrointestinal tract excluding anorectal anomalies has been reported as ranging from 6.8 to 22.5 percent (1-4). Between 1 to 10 percent of cases with esophageal atresia and tracheoesophageal fistula develop hypertrophic pyloric stenosis (HPS) (2-6). Hypertrophic pyloric stenosis does not become clinically evident until the third week or later. At the Göztepe Social Security and Marmara University Hospitals between the years 1983 and 1989 one of the 24 cases of esophageal atresia was complicated by the development of hypertrophic pyloric stenosis. This case forms the basis of this report.

CASE REPORT
The patient T.M. was a male infant born on September 8th 1985 weighing 2.7 kg. Respiratory distress and excess mucus became apparent shortly after birth. Following the diagnosis of EA and distal TEF, on September 11th, the ligation of fistula and a primary anastomosis with a transanastomotic tube were performed through a retropleural approach. The infant tolerated feedings through the transanastomotic tube on the second postoperative day and oral feeding on the sixth day.

The patient was discharged on September 30th. When the baby was 47 days old he was admitted again with feeding difficulties. There had been no weight gain and he was vomiting intermittently. Upper GI series was suggestive of hypertrophic pyloric stenosis and a palpable olive was noted on physical examination. Pyloromyotomy was performed and the patient recovered uneventfully.

DISCUSSION
Most anomalies associated with EA and TEF are apparent in the neonatal period. However HPS is not clinically evident during the first few days of life. The late presence of HPS after 3 weeks of age is often obscured, because the symptom of vomiting is usually attributed to the complications of prior esophageal surgery; such as anastomotic stricture gastroesophageal reflux or esophageal incoordination (5,6)

The etiology of HPS is unknown. Benson assumed pylorospasm as a initiating factor, followed by mucosal and submucosal edema narrowing of the pyloric canal and secondary hypertrophy of pyloric musculature (6). Gastrostomy alone or with transpyloric feeding has also been incriminated as the cause of pyloric stenosis (7,8). It's believed that the continuous gastric decompression reduces intraluminal pressure to a level insufficient to keep the pyloric canal open (9). Since we haven't performed gastrostomy in our case it's hardly acceptable that the gastrostomy plays a role in the development of pyloric stenosis. Similiar cases have appeared in the literature (5-8).

High incidence of HPS in EA patients in comparasion to general population indicates a significant relationship between the two anomalies. Etiological explanation of this association should be by genetic basis rather than mechanical.

HPS should be considered in the evaluation of vomiting in an infant who has had recent surgery for EA and TEF. If an early diagnosis is to be made, in addition to evaluating the esophageal motility the stomach, pylorus and duodenum should be studied for HPS.

REFERENCES

1. *Raffensperger J, Gastrointestinal-tract defects associated with esophageal atresia and tracheoesophageal fistula. J Arch Surg 1970; 101: 241-243.*
2. *Louhimo I, Esophageal Atresia. Primary result of 500 consequtively treated patients. J Ped Surg 1983; 18: 217-229.*
3. *Buker RH, Cox WA, Pauling FW, et al. Complications of congenital tracheoesophageal atresia. Am J Surg 1972; 124: 705-710.*
4. *Andassy RJ, Mahour GH. Gastrointestinal anomalies associated with esophageal atresia or tracheoesophageal fistula. Arch Surg 1979; 114: 1125-1128.*
5. *Franken EA, Saldino RM. Hypertrophic pyloric stenosis complicating esophageal atresia and tracheoesophageal fistula. Am J Surg 1969; 117: 647-649.*
6. *Glasson MJ, Bandrevics V, Cohen DH. Hypertrophic pyloric stenosis complicating esophageal atresia. Surgery 1973; 74: 530-535.*
7. *Ahmed S. Infantilepyloric stenosis associated with major anomalies of the alimentary tract. J Ped Surg 1970; 5: 660-666.*
8. *Czernik J, Raine PAM. Esophageal atresia and pyloric stenosis an association.*
9. *Scharli A, Sieber WK, Kieswetter WB. Hypertrophic pyloric stenosis at the Children's Hospital of Pittsburgh from 1922-1969: A critical review of current problems and complications. J Ped Surg 1969; 4: 108-114.*